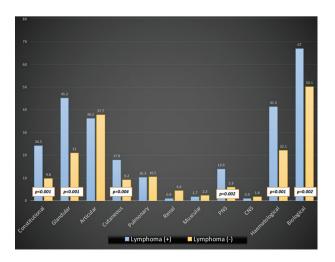
## OP0126 LYMPHOMA ARISING AT THE TIME OF DIAGNOSIS OF PRIMARY SJÖGREN SYNDROME: A HIGHLY-ACTIVE SYSTEMIC SUBSET OF THE DISEASE

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**Objectives:** To analyse the phenotype of patients with primary Sjogren syndrome (SjS) in whom a lymphoproliferative disease is diagnosed concomitantly. **Methods:** By January 2019, The Big Data Sjögren Project included 11,420 consecutive patients with primary SjS recruited from 24 countries of the five

continents. **Results:** 117 (1%) patients were diagnosed with lymphoma and primary SjS synchronously. Age-gender adjusted multivariate analysis identified the following features associated with lymphoma (OR; CI95%): male gender (4.61; 2.88-7.18), White ethnicity (3.51; 1.78-7.91), abnormal oral tests (3.4; 1.38-10.88), positive biopsy (3.2; 1.3-10.17), positive RF (2.27; 1.48-3.53), hypocomplementemia (3.39; 2.06-5.54), and cryoglobulins (4.74; 2.57-8.38). Activity (score > 1) in the constitutional (2.97; 1.86-4.62), glandular (3.11; 2.1-4.57), cutaneous (2.17; 1.28-3.52), peripheral nerve (2.56; 1.4-4.41) and hematological (2.49; 1.64-3.75) ESSDAI domains was associated with lymphoma (frequencies summarized in the Figure).



Conclusion: Patients diagnosed concomitantly with primary SjS and lymphoma have a very specific, highly-active phenotype (men, White, severe oral

involvement, cryoglobulinemic-related immunological markers, and high systemic activity).

Disclosure of Interests: : Soledad Retamozo: None declared, Nihan Acar-Denizli: None declared, Wan Fai Ng: None declared, Antónia Szántó: None declared, Astrid Rasmussen: None declared, Raphaèle Seror Grant/research support from: Pfizer, Consultant for: Bristol-Myers Squibb, Pfizer, Amgen, Eli Lilly, Roche, Celgene, GlaxoSmithKline, MedImmune, Xiaomei Li: None declared, Chiara Baldini: None declared, Jacques-Eric Gottenberg Grant/research support from: Bristol-Myers Squibb, Grant/research support from: Bristol-Myers Squibb, Consultant for: Bristol-Myers Squibb, Lilly, Pfizer, Sanofi-Genzyme, UCB Pharma, Consultant for: Bristol-Myers Squibb, Eli Lilly, UCB, Sanofi-Genzyme, Pfizer, Pulukool Sandhya: None declared, Luca Quartuccio: None declared, Roberta Priori: None declared, Gabriela Hernandez-Molina: None declared, Berkan Armagan: None declared, Aike A. Kruize: None declared, Seung-Ki Kwok: None declared, Marika Kvarnstrom: None declared, Sonja Praprotnik: None declared, Damien Sene: None declared, Roser Solans-Laqué: None declared, Maureen Rischmueller Consultant for: Abbvie, Bristol-Meyer-Squibb, Celgene, Glaxo Smith Kline, Hospira, Janssen Cilag, MSD, Novartis, Pfizer, Roche, Sanofi, UCB, Thomas Mandl: None declared, Yasunori Suzuki: None declared, David Isenberg: None declared, Valeria Valim: None declared, Agata Sebastian: None declared, Gunnel Nordmark: None declared, Hendrika Bootsma: None declared, Hideki Nakamura: None declared, Roberto Giacomelli Grant/research support from: Pfizer, Actelion, Speakers bureau: Actelion, Bristol-Myers Squibb, Merck Sharp & Dohme, Abbvie, Pfizer, Sobi, Roche, Valerie Devauchelle-Pensec Grant/research support from: Roche-Chugai, Speakers bureau: MSD, BMS, UCB, Roche, Benedikt Hofauer Consultant for: Consultant for Galvani Bioelectronics for the area of sleep disorders., Michele Bombardieri Grant/research support from: Celgene, Consultant for: Medimmune, Virginia Fernandes Moça Trevisani: None declared, Daniel Hammenfors: None declared, Sandra Pasoto: None declared, Tamer A Gheita: None declared, Fabiola Atzeni: None declared, Jacques Morel: None declared, Cristina Vollenveider: None declared, Sandra Consani-Fernández: None declared, Xavier Mariette Grant/research support from: Servier, Consultant for: AstraZeneca, Bristol-Myers Squibb, GlaxoSmithKline, Janssen, Pfizer, UCB Pharma, Manuel Ramos-Casals: None declared, Pilar Brito-Zerón: None declared, Elena Bartoloni Bocci: None declared

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## OP0127 DISEASE PRESENTATION OF 1,312 CHILDHOOD-ONSET SYSTEMIC LUPUS ERYTHEMATOSUS: INFLUENCE OF ETHNICITY

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**Background:** To our knowledge the influence of ethnic background in childhoodonset SLE (cSLE) presentation was not evaluated in a large population of Latin American country.

**Objectives:** To assess demographic data, clinical manifestations, laboratory abnormalities and disease activity score in cSLE patients according to ethnic groups at diagnosis

**Methods:** This multicenter study included cSLE patients(ACR criteria) followed in 27 Pediatric Rheumatology services of Brazil. Ethnicities were classified in four groups according to the parents' and all four grandparents' self-reported ethnicity. The statistical analysis was performed using the Bonferroni's correction (p<0.0027).

**Results:** According to ethnic groups, 1,537 cSLE patients were classified in: Caucasian (n=786), African-Latin American(n=526), Asian(n=8) and others/unknown (n=217). Comparisons between 1,312 African-Latin American and Caucasian revealed similar median age at cSLE diagnosis[12.2(2.6-18) vs. 12.1(0.3-18) years,p=0.234], time interval to diagnosis[0.25(0-12) vs. 0.3(0-10) years,p=0.034] and SLEDAI-2K score[14(0-55) vs. 14(0-63),p=0.781] in both groups. The mean number of diagnostic criteria according to SLICC(6.47±1.911 vs. 5.81±1.631, p<0.0001) and frequencies of maculopapular lupus rash (8% vs. 3%, p<0.0001), palate oral ulcers(17% vs. 11%,p=0.001), tongue oral ulcers (4% vs. 1%, p=0.001) and nonscarring alopecia(29% vs. 16%,p<0.0001) were significantly higher in African-Latin American, whereas malar rash(45% vs. 58%,p<0.0001) was more frequent in Caucasian. The presence of antiphospholipid antibody(23% vs. 12%,p<0.0001), low complement levels(58% vs. 41%, p<0.0001) and isolated direct Coombs test (10% vs. 5%,p=0.001) were also significantly higher in the former aroup.

Conclusion: Our study demonstrated that disease presentation severity of African-Latin American cSLE patients is comparable to Caucasian. Mucocutaneous manifestations and autoantibodies profile were the only distinctive features of the former group. The unique mixed background of Brazilian patients probably minimized race diversity spectrum of these patients.

Disclosure of Interests: None declared

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## OP0128 SYSTEMIC LUPUS ERYTHEMATOSUS DISEASE CHARACTERISTICS ASSOCIATED WITH THE TYPE I INTERFERON GENE SIGNATURE: BASELINE DATA OF THE SLE PROSPECTIVE OBSERVATIONAL COHORT STUDY (SPOCS)

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Background: Despite accumulating data on the role of type I interferons (IFN) in the pathogenesis of SLE, real-world, longitudinal clinical data on the type I IFN gene signature (IFNGS) collected from patients with SLE are limited.

Objectives: This initial analysis of the SLE Prospective Observational Cohort Study (SPOCS; NCT03189875) examined the prevalence of the type I IFNGS (high vs low) and its association with baseline SLE disease characteristics in patients with moderately to severely active SLE receiving standard-of-care treatment.

Methods: SPOCS is an international, multicenter, prospective observational cohort study of patients enrolled with moderately to severely active SLE (SLEDAI-2K ≥6 at entry) from Australia, Canada, France, Germany, Italy, Spain, the United Kingdom, and the United States; a completion date of 2022 is planned. Patients

Baseline Demographics and Clinical Characteristics Total				
Variable	population (N=307)	IFNGS-high (N=210)	IFNGS-low (N=83)	<i>P</i> -value <sup>4</sup>
Age (years), median (min-max)	46.0 (18-88)	42.5 (18-82)	50.0 (19-88)	< 0.0001
18-29, n (%)	45 (14.7)	38 (18.1)	5 (6.0)	
30-39, n (%)	71 (23.1)	61 (29.0)	9 (10.8)	
40-49, n (%)	68 (22.1)	37 (17.6)	26 (31.3)	
50-59, n (%)	70 (22.8)	42 (20.0)	23 (27.7)	
≥60, n (%)	53 (17.3)	32 (15.2)	20 (24.1)	
BMI (kg/m <sup>2</sup> ), median (min-max)	26.8 (17-58)	25.5 (17-52)	28.9 (17-58)	0.0029
Age at SLE diagnosis (years), median (min-max)	34 (8–76)	30 (8–76)	42 (8-73)	< 0.0001
Age at first SLE manifestation (years), median (min-max)	31.0 (5-75)	28.5 (7-75)	37.0 (5-66)	0.0005
SLEDAI-2K, total score, median (min-max)	8.0 (4–29)	8.0 (4–29)	6.0 (4–26)	0.0002
SDI, median (min-max)	1.0 (0-10)	1.0 (0-10)	1.0 (0-5)	0.8502
PtGA VAS score, median (min-max) Medical history, n (%)	50.0 (0-100)	48.5 (0-100)	58.5 (5-98)	0.0131
Any comorbidity	254 (82.7)	167 (79.5)	76 (91.6)	0.0136
CNS disorders	28 (9.1)	24 (11.4)	1 (1.2)	0.0048
CV risk factors and CBV disease	101 (32.9)	60 (28.6)	36 (43.4)	0.0150
Joint disease (SLE and non-SLE)	196 (63.8)	123 (58.6)	65 (78.3)	0.0015
Diabetes mellitus	28 (9.1)	14 (6.7)	13 (15.7)	0.0164
Laboratory tests	. ,	. ,	. ,	
Complement C3, (mg/dL)	96.5 (31-191),	90.5 (31-191),	104.0 (60-171),	0.0026
median (min-max), n	142	100	35	
Complement C4, (mg/dL)	17.0 (2-177),	14.0 (2-177),	21.0 (8-164),	0.0005
median (min-max), n	138	97	34	
ANA positive, n (%)b	37 (75.5)	30 (93.8)	6 (42.9)	0.0004
Sm, RNP autoantibody positive, n (%)b	12 (36.4)	12 (50.0)	0 (0.0)	0.0261
dsDNA antibody positive, n (%)b	15 (26.8)	14 (37.8)	1 (7.1)	0.0411

 dsDNA antibody positive, n(%)<sup>b</sup>
 15 (26.8)
 14 (37.8)
 1(7.1)
 0.0411

 ANA, antinuclear antibody; BMI, body mass index; CBV, cerebrovascular; CNS, central nervous system; CV, cardiovascular; dsDNA, double-stranded deoxyribonucleic acid; IFNGS, interferon gene signature; max, maximum; min, minimum; PiGA, Pattent Global Assessment of Disease Activity; SDI, Systemic Lupus International Collaborating Clinics (SLICC)/American College of Rheumatology (ACR) Damage Index; SLEDAI-2K, Systemic Lupus Erythematosus Disease Activity Index-2000; Sm, Smith antibody; RNP, ribonucleoprotein antibody; VAS, visual analog scale.

 PIRGS-light vis FINGS-10w, determined by chi-square test, Mann-Whittey U test, or Fisher exact test.
 <sup>1</sup>Total number of patients tested (total, high, low): ANA: 49, 32, 14; Sm, RNP; 32, 24, 6; dsDNA: 56, 37, 14.

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are evaluated biannually during a 3-year follow-up period. At each visit, data are collected on disease activity and damage, treatment received, type I IFNGS (stratified high or low IFNGS based on a predefined cutoff), and several patientreported outcomes.

Results: As of November 15, 2018, a total of 307 patients were enrolled in SPOCS (North America, n=184; Europe, n=123), of whom 96.1% (n=295) were female, with a median age of 46 years (range: 18-88). At study entry, the prevalence of high type I IFNGS was 70.5% (n=210) vs 27.9% (n=83) for low type I IFNGS, with 1.7% (n=5) unknown (table, 9 missing data). IFNGS-high patients were younger than IFNGS-low patients (median: 42.5 years [range: 18-82] vs 50 [19-88], P<0.0001) and diagnosed with SLE at an earlier age (P<0.0001). SLE-DAI-2K scores were greater for IFNGS-high patients vs IFNGS-low patients (P=0.0002), while SDI scores were similar between the two groups. Fewer comorbidities were reported for IFNGS-high patients than for IFNGS-low patients (79.5% [n=167] vs 91.6% [n=76], P=0.0136). Lower complement C3 and C4 levels were observed in IFNGS-high vs -low patients. At study entry, antinuclear antibodies and ribonucleoprotein antibodies were more frequent in the IFNGS-high vs IFNGS-low subset of patients. A greater percentage of IFNGS-high patients were dsDNA antibody positive vs IFNGS-low patients (P=0.0411).

Conclusion: The profile of patients with a baseline high type I IFNGS differed from those with a low type I IFNGS, in that those with a high type I IFNGS comprised a group that were on average younger, had greater SLEDAI-2K scores, were more serologically active, and seemed to have fewer comorbidities. As the lupus community evolves from using a classical clinical classification of patients to one based on molecular signatures, it is important to understand the role of the type I interferon pathway on disease activity, treatment, and outcomes. SPOCS recruitment and follow-up are ongoing.

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OP0129

## CAPS CRITERIA FAIL TO IDENTIFY MOST SEVERE PATIENTS WITH ANTIPHOSPHOLIPID SYNDROME ADMITTED TO THE INTENSIVE CARE UNITWITH A NEW THROMBOTIC MANIFESTATION

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Background: Catastrophic antiphospholipid syndrome (CAPS) is the most severe manifestation of antiphospholipid syndrome (APS), characterized by the simultaneous occurrence of thrombosis in multiple organs.

Objectives: The objectives of this study were to evaluate the distribution and the prognosis of CAPS criteria in APS patients admitted to the intensive care unit (ICU) with acute thrombotic manifestation.

Methods: We conducted a multicentre retrospective study, from January 2000 to September 2018, including all APS patient admitted to 24 French ICUs with any new thrombotic (arterial, venous or microvascular) manifestation.

Results: 134 single patients were admitted to the ICU for 152 episodes. The number of patients with definite CAPS, probable CAPS and no CAPS was: 11 (7.2%), 60 (39.5%) and 81 (53.5%) respectively. We compared patients with definite/probable CAPS (group 1, n=61) and no CAPS (group 2, n=73). General characteristics and APS-involved organs are reported in Table 1. APS characteristics and biological findings before admission were comparable between both groups. In-ICU and in-hospital length of stay, day-0 SAPS II and day-0 SOFA in-ICU severity scores